Multiple Sebaceous Tumors May Show Syndrome

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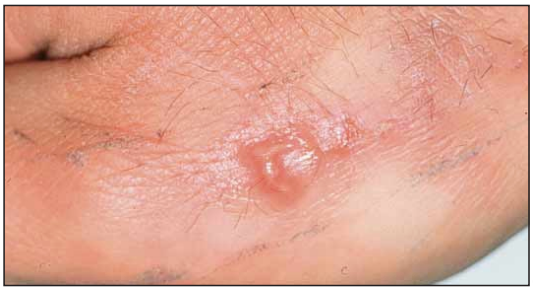

COURTESY DR. ISTVAN JUHASZ

BY NANCY WALSH
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BUDAPEST, HUNGARY — A 56-year-old man with a 7-year history of multiple small cutaneous tumors histologically identified as basal cell carcinomas presented with new skin-colored tumors on his forehead and back, K. Gaspar, M.D., said at an international symposium sponsored by the European Academy of Dermatology and Venereology.

Family history revealed that his brother, who was 2 years older, had undergone a hemicolectomy for adenocarcinoma several years earlier. The brother was called in for examination and was found to have a number of tiny, crust-covered tumors on his back and face that resembled sebaceous gland tumors.

Both of his sisters had had surgery for gynecologic cancers; one had died 2 years earlier of ovarian cancer. His mother also had died of an undetermined gynecologic cancer.

After excision, the patient's new tumors were found to be sebaceous adenomas, a result that raised the possibility that this patient and his family members had Muir-Torre syndrome, said Dr. Gaspar of the department of dermatology at Medical and Health Science Center, University of Debrecen (Hungary).

Muir-Torre syndrome is a rare genodermatosis that exhibits an autosomal dominant inheritance pattern. The syndrome, which is characterized by sebaceous lesions and visceral malignancies, is considered a phenotypical variant of hereditary nonpolyposis colorectal cancer. Both conditions are caused by inherited DNA mismatch repair defects, most commonly on the *MSH2* and *MLH1* genes.

Among the skin lesions characteristic of Muir-Torre syndrome are sebaceous adenomas, epitheliomas, and carcinomas. Adenomas appear as skin-colored or yellowish papules or nodules, sometimes with a central depression. Histologic evaluation in this case showed orthokeratotic, dilated follicles, numerous nucleoli, and a lack of sebum.

Sebaceous carcinoma appears as a poorly demarcated, asymmetrical, solid tumor with an irregular border. Histology showed that this lesion had deep penetrating tumor cell nests that obliterated the normal structure of the sebaceous gland.

This case illustrates the importance of a careful search for possibly asymptomatic abdominal tumors in the setting of multiple sebaceous tumors. Patients whose cutaneous tumor cells are genetically unstable (characterized by mutations of short DNA sequences known as microsatellites) also are at heightened risk for developing subsequent abdominal cancers. "Therefore, microsatellite screening can play a significant role in cancer prevention," Dr. Gaspar said.

In more than half of cases the visceral malignancy is colorectal carcinoma. Genitourinary, hematologic, and head and neck sites also have been reported.

Vigilance is necessary, and repeated surgeries for both skin and visceral tumors can be expected. Family members also must be evaluated and followed. But with careful follow-up, the prognosis of Muir-Torre syndrome is good, with 10-year survival exceeding 50%, Dr. Gaspar said at the symposium, also sponsored by the Hungarian Dermatological Society.

At the patient's most recent follow-up examination, his tumor markers were normal, but radiographic evaluation revealed metastases to the paraaortic lymph nodes. He was started on a chemotherapeutic regimen that included irinotecan. ■